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Case report

Subcorneal pustular dermatosis in a 7-year old Saudi child: A case report and review of the literature☆

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Abstract

Subcorneal pustular dermatosis (SCPD) also known as Sneddon–Wilkinson disease (Sneddon and Wilkinson, 1956) is a rare, benign, chronic, sterile pustular eruption which usually develops in middle-age or elderly women; it is rarely seen in childhood and adolescence (Johnson and Cripps, 1974). The primary lesions are pea-sized pustules classically described as half-pustular, half-clear flaccid blisters. Histologically the most important feature is a subcorneal accumulation of neutrophils with the absence of spongiosis or acantholysis. In this paper we present the case of a 7-years-old boy diagnosed with SCPD based on the characteristic clinical and histological features. Oral and topical corticosteroid has been successfully used in the treatment of the disease.

Keywords: Subcorneal pustular dermatosis (Sneddon–Wilkinson disease); Histopathology; Immunofluorenscence; Immunoglobulin A; Dapsone; Prednisolone; Clobetasone proprionate

1. Case report

A 7-year-old boy was admitted to our clinic with a recurrent itchy pustular eruption located on the trunk on and off in the last six months (Figs. 1 and 2). The palms, soles, and mucous membrane were spared, and no lymphadenopathy or hepato-splenomegaly was present. There were no abnormalities of the nails and mucous membranes.

A complete blood count and the studies of serum biochemistry showed normal results; moreover serum protein electrophoresis had negative results. Normal Thyroid hormone profile reads Rheumatoid factor as negative.

The dermatologic examination revealed multiple-grouped flaccid pustules varying in size and some of them tended to coalesce to form annular pattern and superficial crusts on the normal or mildly erythematous skin of trunk and upper extremities. Healed lesions presented as residual hyperpigmentation and new lesions in the periphery.

Histopathology demonstrated a subcorneal vesiculobullous dermatitis (Fig. 3); the pustule is located immediately below the stratum corneum and contains mainly neutrophils with few eosinophils. The underlying epidermis to the pustule shows slight intercellular edema. In the dermis, superficial blood vessels are surrounded by a nonspecific mixed inflammatory cell infiltrate consisting of neutrophils and mononuclear cells. Direct immunofluorescence studies

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☆ We describe a rare case of Subcorneal pustular dermatosis (SNEDDON–WILKINSON DISEASE) in a 7-year old Saudi boy presented to the outpatient clinic in King Fahad Hofuf Hospital. We highlight the clinical features and histopathological findings that distinguish this case from other pustular diseases. We also describe the management and the outcome of this case.

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Figure 1. A recurrent itchy pustular eruption located on the trunk.

Figure 2. Closer view.

are negative for immunoglobulin A (IgA) intercellular accumulation. On the basis of this finding, associated to histopathological features and the clinical date, a diagnosis of subcorneal pustular dermatitis (SCPD, Sneddon–Wilkinson disease) was made.

With treatment in the form of tapering dose of prednisolone starting with 30 mg over three weeks' time and then topical clobetasone proprionate the patient showed great improvement within eight weeks. The patient was lost to follow-up but presented after 6 months upon relapse. The same course of treatment was repeated with significant improvement within two months. No further follow up of patient could be accomplished.

2. Discussion

Subcorneal pustular dermatosis is a chronic, relapsing, pustular eruption, generally involving the trunk, which affects mainly women over 40 years of age according to Sneddon and Wilkinson's original report (Sneddon and Wilkinson, 1956).

Children can have various bullous and pustular skin diseases like psoriasis, pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid as well as dermatitis herpetiformis; all of these were once thought to be unique to people in the fourth-fifth decade of life. Subcorneal pustular dermatosis appears to be another one of these diseases (Scalvenzi and Palmisano, 2013).

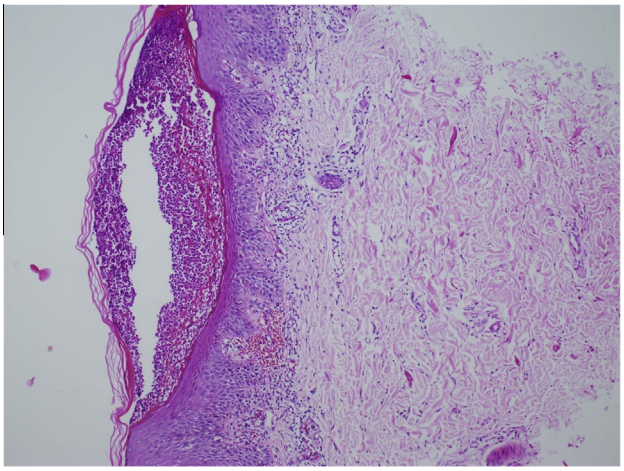

Figure 3. Histopathology demonstrated a subcorneal vesiculo-bullous dermatitis (Fig. 3); the pustule is located immediately below the stratum corneum and contains mainly neutrophils with few eosinophils.

Only 15 cases of pediatric SCPD are described in the literature (Johnson and Cripps, 1974; Yayli et al., 2006).

Even if SCPD is an uncommon condition in childhood, it must be considered as a possible cause of sterile pustular eruptions in a child. An accurate physical examination, a complete blood count, and studies of serum biochemistry are strongly recommended to exclude a pathology in association.

The etiopathogenesis of SCPD is not well known. Culture of the pustules is sterile. A relationship with Pyoderma gangrenosum (Scerri et al., 1994; Marsden and Millard, 1986), benign monoclonal IgA gammopathy (Kasha and Epinette, 1988; Scerri et al., 1994), IgA myeloma (Atukorala et al., 1993; Takata et al., 1994; Vaccaro et al., 1999), SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome (Scarpa et al., 1997), Crohn's disease (Delaport et al., 1992), Rheumatoid arthritis (Butt and Burge, 1995), and Hyperthyroidism (Taniguchi et al., 1995) has been documented.

In our case, the history, physical examination, and laboratory results did not reveal any systemic associations. Moreover some cases, which were consistent with SCPD according to the clinical and histologic features, have been reported with the presence of an intercellular IgA deposition within the epidermis (Hashimoto et al., 1987).

This disease involves more frequently the trunk as in this case. Other sites can be involved like the intertriginous areas, and flexor aspects of the limbs; more rarely the face is implicated. Pustules on palms and soles have also been reported (Takematsu and Tagami, 1993), while mucous membranes are almost never affected.

The differential diagnosis of SCPD includes impetigo, pustular psoriasis, dermatophyte infection, immunobullous

diseases (dermatitis herpetiformis, pemphigus, linear IgA disease, and intercellular IgA diseases) and acute generalized exanthematous pustulosis (Roujeau et al., 1991). Pathogenic organisms are cultured from pustules in impetigo and the condition responds to antibiotics.

Pustular psoriasis, either of the acute von Zumbusch type with small pustules or the spreading annular type, may resemble subcorneal pustular dermatosis (Burge, 2010). Spongiosis is not a feature of subcorneal pustular dermatosis, but spongiform pustules, which are an integral part of the epidermis, are characteristic of pustular psoriasis (Wolff, 1981; Sanchez et al., 1983). Subcorneal pustular dermatosis, unlike pustular psoriasis, responds to dapsone. Some authors consider that subcorneal pustular dermatosis is part of the spectrum of pustular psoriasis (Sanchez et al., 1983). Acute generalized exanthematic pustulosis is distinguished by its acute onset in a febrile patient with a history of exposure to a candidate drug. The histology shows spongiform pustules (Todd et al., 1991). A dermatophyte infection can be easily excluded with a direct microscopic examination of fungal elements. IgA deposition in the dermal papillae distinguishes SCPD from dermatitis herpetiformis.

Biopsies from early lesions show a perivascular inflammatory infiltrate with neutrophils and occasional eosinophils. Neutrophils migrate through the epidermis, without forming spongiform pustules, to collect beneath the stratum corneum in subcorneal pustules. The pustules sit on the surface of the epidermis rather than being an integral part of it. A few acantholytic cells may be found in old lesions. Ultrastructural studies show cytolysis of single cells and invasion by neutrophils. Both direct and indirect immunofluorescent studies are negative in classical cases, but recently some cases have been described with intercellular IgA within the epidermis. The relationship between the intercellular IgA dermatosis and subcorneal pustular dermatosis is not clear.

Dapsone remains the treatment of choice but its safety is still debatable especially in childhood and a close follow-up is required; the minimal effective dose to suppress the disease should be determined in these patients. Sulfapyridine (1.0–3.0 g daily) is also beneficial; in our patient systemic corticosteroids were given and shown to be effective along with topical. Etretinate (Todd et al., 1991; Szabo and Hamm, 1992; Vaccaro et al., 1999) and acitretin (Marlière et al., 1999; Yayli et al., 2006) have been used. Isotretinoin was found ineffective at a dose of 0.5 mg/kg/d (Rutman et al., 1988). Broad-band UVB (Park et al., 1986), narrow-band UVB (Orton and George, 1997; Cameron and Dawe, 1997), PUVA (Todd et al., 1991; Bauwens et al., 1999) and Re-PUVA have also been reported as effective. Colchicine (Kawaguchi et al., 2000) and topical tacalcitol (Hashimoto et al., 1987) have been recommended. There are individual case reports of the use of etanercept (Bedi, 2007), combination of adalimumab and mycophenolate mofetil (Howell et al., 2005) and infliximab, which produced rapid but short-lived benefit (Bonifati et al., 2005). In cases associated with myeloma, the skin lesions may improve when the paraprotein is reduced by chemotherapy (Takata et al., 1994).

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